Headache

Introduction

There are three headache subtypes: primary headaches, secondary headaches, and painful cranial neuralgias.

Primary headache disorders include migraines, tension-type headaches, and cluster headaches. Cluster headaches are just one type of trigeminal autonomic cephalalgia, but at this stage in your training, we are reducing those to just cluster headaches. These headaches are not life-threatening and do not warrant imaging. Primary headaches, which are caused by a specific headache disorder, are more commonly seen in practice and are defined by clinical criteria.

Secondary headaches result from another underlying process and are defined by such. These headaches may be associated with significant morbidity and mortality and require early identification—image early if red flags are present. Patients with serious secondary headaches typically have acute or subacute symptoms and a headache pattern that is progressive or unstable.

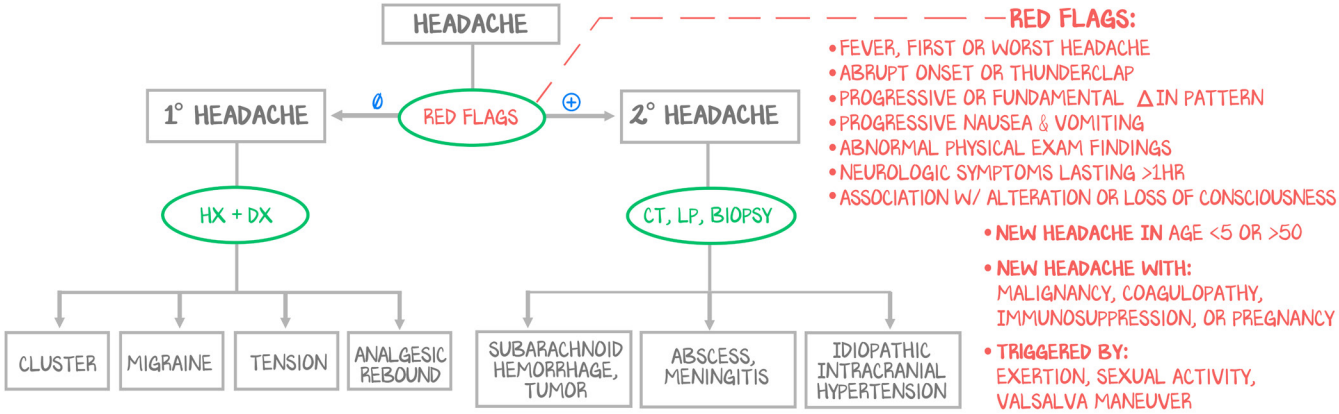

Figure 5.1: Identifying Headaches as Primary or Secondary
If there are red flags, a secondary headache is more likely, and imaging is required. If there is fever, focal neurologic deficit, or papilledema, urgent imaging is required. If there are other red flags alone, imaging is still indicated, but electively. If there are no red flags, entertain the primary causes of headaches.

Painful cranial neuralgias are face pain associated with any cranial nerve. We are going to teach you one—**trigeminal neuralgia**. Like cluster headaches, there are many types and subtypes. For this stage of your training, you should learn the headache diagnoses—cluster, migraine, and tension—and the one face pain diagnosis—trigeminal neuralgia.

A word of caution. We are not teaching you the trigeminal autonomic cephalgias (the subtypes of cluster headaches) because the umbrella diagnosis has "trigeminal" in its name (erroneously leading learners to believe that it is the umbrella for trigeminal neuralgia) and because that level of detail is too minute. Likewise, we are not teaching you the painful cranial neuralgias (the subtypes of trigeminal neuralgia) because they are uncommon, both in life and on licensing examinations.

So, this lesson includes cluster headaches, migraines, tension headaches, trigeminal neuralgia, and secondary headaches.

Primary 1: "Cluster Headaches"

Because the mechanism behind cluster headaches is **not well elucidated**, you cannot be expected to know it. Instead, discussing cluster headaches requires more of a clinical approach than a mechanistic one—recognize the headache syndrome, treat accordingly.

Cluster headaches are of **short duration** but present with **extreme pain**. The patient will describe **ten-out-of-ten unilateral pain**, most often located intra- or **supraorbitally**. Patients must also have at **least one autonomic symptom ipsilateral** to the pain (conjunctival injection, rhinorrhea, lacrimation, nasal congestion, or Horner syndrome). Unlike patients with migraines (below), these patients are **unlikely to hold still**, pacing about trying to find relief. Attacks typically last anywhere from **15 minutes to 3 hours**. They can occur up to eight times per day, although patients typically experience attacks twice per day, usually at night. Most patients will have attacks for weeks to months and then have a remission for months up to years. **Men** are much more likely to experience cluster headaches than women**,** whereas all other primary headache syndromes are more prevalent in women.

Complete transection of the trigeminal nerve does **not prevent symptoms**. Thus, cluster headaches are likely to be caused by a brainstem or thalamic lesion. The patient **senses pain**, but no cause for the pain is found. EEGs have not revealed seizure patterns. However, it stands to reason that if there were seizure activity, it might be so focal and deep in the brain that the seizure is not perceived by the electrodes on the scalp. To be frank, this is pure speculation, as the pathogenesis is currently idiopathic (aka, we have no idea). Treatment with **100% O_2** is abortive. Nearly two-thirds of attacks are aborted by oxygen. For those that don't respond to oxygen, **triptans** (subcutaneous sumatriptan or intranasal zolmitriptan or sumatriptan) are used for acute refractory attacks.

Verapamil is the preferred agent for prophylaxis. Don't be fooled. This is an L-type calcium-channel blocker that slows the heart rate. This is not an arterial vasodilatory like the dihydropyridine calcium-channel blockers ("-dipines"), so verapamil most likely alters signal transduction within the sensory tracts.

	CLUSTER HEADACHES	CPH	SUNCT
Duration	15–300 minutes	2–30 minutes	1–600 seconds
Frequency	8/day	40/day	100/day

Table 5.1: Trigeminal Autonomic Cephalalgia
Don't memorize this table. It's here so that you can see the inverse relationship between duration and frequency. This is for exposure's sake only. We said we wouldn't teach it to you, but you should be exposed to it. CPH, chronic paroxysmal hemicrania; SUNCT, short-lasting unilateral neuralgiform headache.

Primary 2: Migraines

We built this curriculum to polarize the diseases that medical science recognizes as separate disease processes and, at the same time, purposefully group diseases together when there are known connections between their disease processes. Medical science has not elucidated the mechanism behind migraines, but migraines have certain symptoms that overlap heavily with those of seizure disorder, especially focal seizures with awareness and sensory symptoms. We didn't teach you that specific syndrome, and we don't want to. You are going to draw parallels between migraines and seizures—auras before they start, a hangover in place of a postictal state, using antiseizure medications as prophylaxis—that, according to medical science, you shouldn't draw. But there is evidence of their connection. Patients with epilepsy are twice as likely to get migraines, and patients with migraines are twice as likely to develop epilepsy. It is almost as if migraines are a very slowed down, less intense version of a seizure. We know a lot about the underlying pathogenesis of seizures. Medical science has not yet elucidated the pathogenesis of migraines. And their treatment is grossly different. We are going to intentionally call out the similarities as they arise to force your brain to consciously see them and then consciously separate them, rather than allowing your subconscious to connect them erroneously. (*We personally think a connection is there that medical science is soon to elucidate.)*

Migraine headaches are not "really bad headaches" that can be treated with "aspirin, acetaminophen, and caffeine." The over-the-counter "migraine relief" or "migraine strength" medications are merely a combination of things that reduce pain and pep you up (aspirin, acetaminophen, and caffeine). If you have had a migraine, or know someone who has migraines, you know what one is. Most people have no idea, and "migraine" is used colloquially to mean "a really bad hangover."

Migraine headaches are **unilateral** and will last **up to three days** if not aborted. **Sleeping** will usually abort the attack. Patients often feel groggy or hungover the day after one breaks (like a slow subacute postictal state). The pain is **throbbing** or pulsatile, of **moderate to severe** intensity, and often associated with autonomic features—**photophobia**, **phonophobia**, and **nausea/vomiting**. The patient will be **lying still** in a dark room, their ears and eyes covered. Some patients experience an **aura**, pre-migraine symptoms that portend the onset of the migraine (like a seizure aura). Migraines often have triggers. Each person's migraine triggers are different, but commonly identified triggers are **menstruation**, **chocolate**, **nitrates,** lack of sleep, and **MSG** (again, similar to seizures).

Did you see how vastly opposite that is from cluster headache? Cluster headaches are short-lived, have no eye or ear symptoms, and the patients scramble to find pain relief.

Acute management for migraines starts with over-the-counter remedies—caffeine, aspirin, and acetaminophen or NSAIDs, such as ibuprofen. If that fails, use **triptans**, the most common of which is sumatriptan. The remedies come in various formulations (nasal spray, pill, subcutaneous injection) and can be tailored to the patient's migraines. Triptans were thought to fix migraines because of vasoconstriction. They do abort migraines, but it is unlikely to be due to their vasoconstrictive effects. Ubrogepant and lasmiditan, medications without vasoconstrictive properties, work well on migraines (they are brand new and not something you should be responsible for knowing). Instead, the **side effects** of triptans may be caused by their vasoconstrictive effects. Patients with **atherosclerotic disease** (especially in the coronary vessels) should not take triptans, as transient vasoconstriction will lead to ischemia. For those patients, there are alternatives that we don't want you to learn (primarily antiemetics and prokinetic agents).

The treatment used to be "triptans and ergot alkaloids." **Ergotamine** is **no longer used**. It is a more potent vasoconstrictor and can lead to ischemia in the digits of patients without atherosclerotic disease. And as we now know that vasoconstriction isn't the mechanism by which migraines are broken, ergotamine is no longer worth giving.

Prophylaxis for migraines is indicated when attacks are frequent, long, and cause severe disability. We did not include the cut-off numbers because you aren't learning checklist medicine. On a licensing exam, the patient will be someone who either OBVIOUSLY does not need prophylaxis or OBVIOUSLY does. There are five medications that have level-A evidence for reducing the frequency and severity of migraines: the β-blockers **propranolol**, timolol (we want you to associate this with glaucoma), and metoprolol (associate this with heart disease), and the broad-spectrum, multiple-mechanism antiepileptic drugs **topiramate** and **valproate**. There is also good evidence for the use of the antidepressant **venlafaxine**.

What we want you to learn right now is that medical science is still figuring out migraines. If there are no other comorbidities, pick **propranolol**. If there is seizure, use either **topiramate or valproate**. And if there is a need for mood stabilization in bipolar disorder, pick **valproate**.

Above all else is **patient behavior**. Regulation of sleep patterns, frequent small meals, adequate hydration, and daily aerobic exercise are all extremely helpful. Regular work and school schedules should be encouraged. Stimulants, such as caffeine and nicotine, must be eliminated or limited. Diet should be modified to avoid additives, such as monosodium glutamate and artificial sweeteners. And above all else, **avoid known triggers**.

Primary 3: Tension

Everyone has experienced tension headaches. That band across the front of your forehead after you drank too much alcohol the night before. The soreness in your shoulders and neck after you've been in the same position studying all day. The headache you get when you're used to three cups of coffee, and you haven't had one by noon. **Rarely do these come to medical attention.** All humans have experienced a tension headache. Almost no one considers them acute enough to seek care, and simply treat them with over-the-counter pain relievers—NSAIDs or acetaminophen.

On licensing examinations, the question will describe a **bilateral band-like** pain across the forehead. There are no warning signs—no photophobia, phonophobia, or focal pain and the pain is not severe. The patient may not want to walk to the cabinet to get their OTC pain relief, but they can. There is no exacerbation with regular activity. Without treatment, the headache will be **mild** in severity and **last 4–6 hours**.

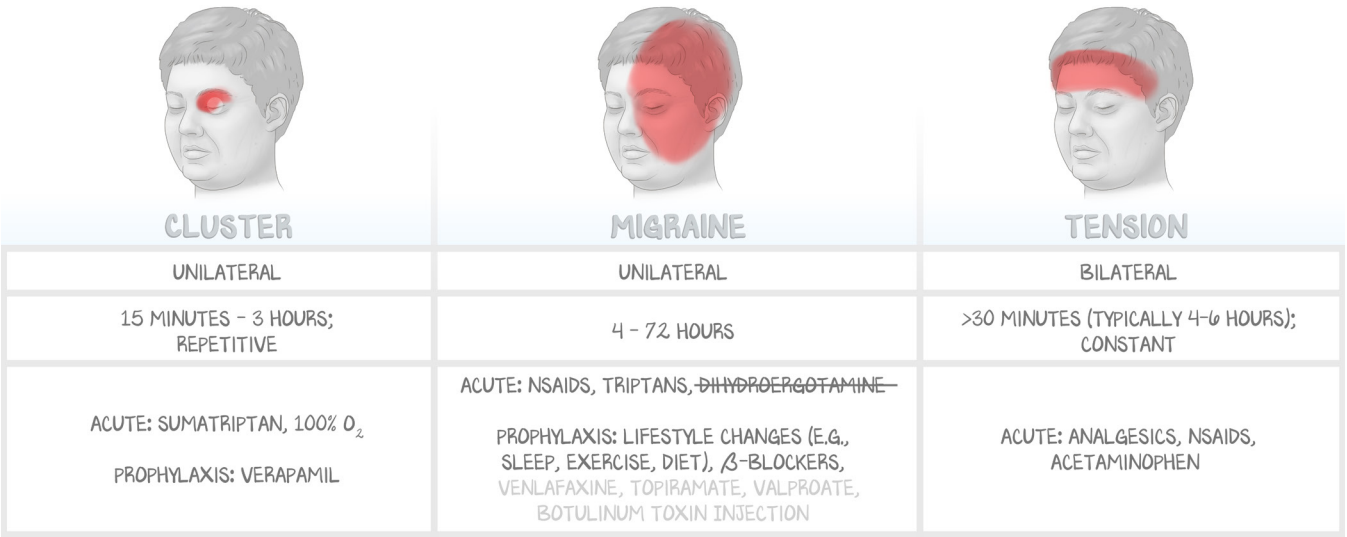

Figure 5.2: Sensory Pain Types
A review of the primary headaches, a summative view of their differences, and their characteristic features.

No Headache, Face Pain = Trigeminal Neuralgia

Trigeminal neuralgia presents nothing like any of the above headache syndromes. But the way it is written in a vignette can trip up novice learners, who may mistake trigeminal neuralgia for cluster headaches or vice versa. Trigeminal neuralgia is a "*seizure of the trigeminal nerve.*" It results in the sudden onset of **unilateral pain**, usually in **one division** of the trigeminal nerve (although it can involve all three). The episodes increase in intensity and frequency over time. Like in cluster headaches, the pain is **severe**, often described as **electric shocks** (stabbing or lancinating pain). This likely represents an overactive sensory syndrome, triggered by stimulating one of the sensory axons in the trigeminal distribution (chewing, talking, or touching the face may set off an attack). They are **brief**—less than a minute per attack. Use **carbamazepine** as prophylaxis.

Secondary Life-Threatening Headaches

This section is about pattern recognition, not pathogenesis. You have already studied brain bleeds, hydrocephalus, brain cancer, increased intracranial pressure, meningitis, etc. This is about recognizing "headache" as part of a syndrome, and recognizing which syndrome the headache is stemming from. All of them require imaging and intervention.

Subarachnoid hemorrhage is described as a **rapid crescendo** ("thunderclap") headache. It is rapid onset (< 1 minute) and classically described as *"the worst headache of my life."* If there were a **sentinel event** (headache a week ago that got better), then this supports subarachnoid hemorrhage, especially on a vignette. Subarachnoid hemorrhage can be diagnosed by CT or **xanthochromia** (old red blood cells on lumbar puncture).

Other **brain bleeds** and **brain cancers** will cause focal neurological deficits and headaches. Any focal neurological deficit with a new headache warrants imaging.

Fever and headache point towards brainflammation—either meningitis, encephalitis, or abscess. When there is bacterial meningitis without abscess, there is usually only fever and headache. With encephalitis (infection of the parenchyma itself, often by a virus), there will be fever, headache, and altered mental status (encephalitis causes encephalopathy). If there is a fungal infection (*Cryptococcus*) or abscess, there will be fever, headache, and seizure OR fever, headache, and focal neurological deficits. The Kernig and Brudzinski sign tests, when performed correctly, are extremely useful if positive. However, often nuchal rigidity is all you are provided.

Headache with papilledema is a sign of increased intracranial pressure. If a patient has a new headache and a good fundic exam demonstrates papilledema, you scan that patient's brain.

Giant cell arteritis (aka **temporal arteritis**, although the name "giant cell" is preferred because it presents with giant cells and can affect an artery other than the temporal artery). The patient will be **old** (> 50 years) and have **ipsilateral jaw claudication**, fever, and **palpable nodular** temporal arteries. Their vision is at risk, and it may present with **monocular vision loss**. There will be an **elevated ESR** and **CRP**. Give **steroids** and take a biopsy to confirm.

There are many others, but essentially, fever and headache, focal neurological deficit and headache, or papilledema with headache warrant imaging.